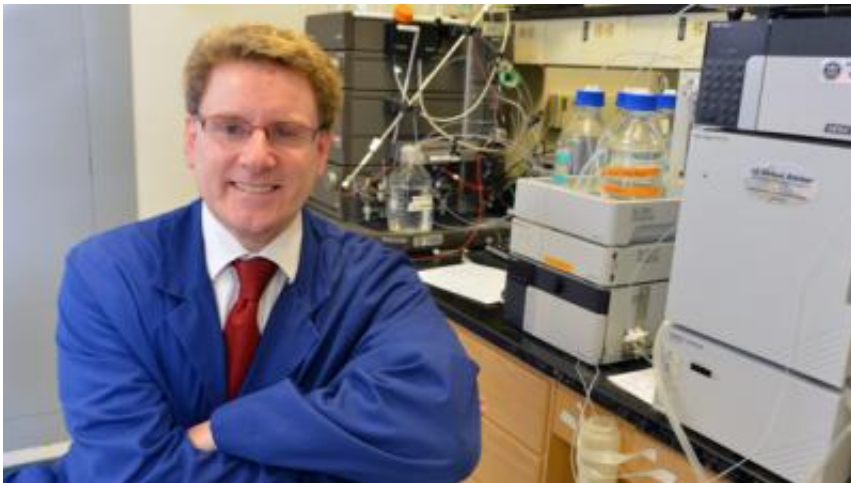


Carnitine supplement may improve survival rates of children with heart defects

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Dr. Stephen M. Black, cell and molecular physiologist at the Vascular Biology Center at the Medical College of Georgia at Georgia Regents University Credit: Phil Jones

A common nutritional supplement may be part of the magic in improving the survival rates of babies born with heart defects, researchers report.

Carnitine, a compound that helps transport fat inside the cell powerhouse where it can be used for [energy production](#), is currently used for purposes ranging from weight loss to chest pain.

New research shows it appears to normalize the blood vessel dysfunction

that can accompany [congenital heart defects](#) and linger even after [corrective surgery](#), said Dr. Stephen M. Black, cell and molecular [physiologist](#) at the Vascular Biology Center at the Medical College of Georgia at Georgia Regents University.

"My hope is this is going to have a major, major impact on survival of babies," Black said. About half the babies born with heart defects have excessive, continuous high pressure on their lungs from misdirected blood flow. Early surgery can prevent full-blown pulmonary vascular disease, but scientists are finding more subtle disruptions in the signaling inside [blood vessels](#) walls that can be problematic – even deadly – up to 72 hours after surgery.

The good news is the changes are reversible and that carnitine speeds recovery and can even prevent the damage in a lamb model of these [human heart](#) defects, according to studies published in the journal *Pediatric Research*.

Normally, most blood flow bypasses the lungs in utero when the placenta provides blood and oxygen for the baby. Baby's first breaths naturally expand the lungs and blood vessels, activating a process inside the lining of vessels that enables them to accommodate the initial blood surge, then reduce pressure quickly, dramatically and permanently.

This natural transition doesn't occur when heart defects misdirect blood flow. "It's kind of like a chronic fetal-to-newborn transition," said Black, the study's corresponding author. Lungs get pounded with about three times the normal flow and, even when surgeries are done as early as possible to repair the defect, correct [blood flow](#) and protect the lungs, the 20 percent death rates from acute pulmonary hypertension have remained unchanged for a decade. "That's 1 in 5 kid (with this condition)," Black said.

Left unchecked, the barrage thickens blood vessels, making them unresponsive, much like those of an elderly individual who has lived for years with uncontrolled high blood pressure. The comparatively brief periods of pounding these babies experience impairs the ability of the endothelial cells, which line blood vessels, to produce nitric oxide, a major dilator of blood vessels.

The shear force disrupts carnitine homeostasis, weakens the mitochondria (the cell powerhouse) and impairs nitric oxide production. To make bad matters worse, the precursor to nitric oxide instead makes more peroxynitrite, prompting endothelial cells to grow and thickening blood vessels. Black was also corresponding author of a recent study in the *Journal of Biological Chemistry* that showed peroxynitrite does this by turning on the cell survival protein kinase Akt1.

The new study indicates that even without fixing the heart defect, high daily doses of carnitine in the first four weeks of life can prevent endothelial dysfunction. In fact, the laboratory lambs' ability to make [nitric oxide](#) is preserved even without the benefit of heart surgery and the responses to the chemical activity that enables blood vessel dilation is normalized, Black said.

Study co-author Dr. Jeffrey Fineman, a whole-animal physiologist and physician at the University of California, San Francisco, developed the model, a lamb whose four-chambered heart is very similar to humans. In utero surgery that misdirects too much blood to the lungs, means that, like children, the lambs are born with the defect.

Black is now working with Fineman, who is pursuing additional funding to resolve questions such as the optimal dosage and timing for giving carnitine. "Do you want to give it for six weeks when you only have to give it for six hours?" Black said. The researchers also plan to examine carnitine homeostasis in the blood of children with [heart defects](#) to see if

it's disrupted. If it is, they plan to start clinical trials.

About 1 in 125 babies are born with a heart defect each year in the United States, according to the March of Dimes. The research was funded by the National Institutes of Health, the Foundation Leducq and the American Heart Association.

Provided by Medical College of Georgia

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